sized musculoelastic arteries and, to a lesser degree, veins. The ratio of male to female patients in fatal cases is 3:1, and most are younger than 2 years of age at the time of death.

Special tests are required to diagnose coronary artery aneurysms, which affect about 20% of patients. Two-dimensional echocardiography is sensitive in detecting left coronary artery aneurysms. Generally, both coronary arteries are aneurysmally dilated in the proximal area. Most of the coronary artery lesions regress in size within a year while the rest have persistent aneurysm or narrowed, tortuous arteries or at least minor permanent damage.

The treatment of Kawasaki disease is supportive care. Until recently administration of high doses of aspirin was the treatment of choice, but this is now undergoing further study. It seems likely that a lower dosage of aspirin will be recommended in the near future. Steroid therapy is contraindicated. In an unpublished study of 300 patients by Drs Kusakawa and Yanagawa of Japan, patients treated with steroid therapy had a significantly higher incidence of aneurysm formation than children treated only with aspirin. Patients with cardiac abnormalities are observed closely during and following their initial illness and should be seen periodically throughout childhood and adolescence for further cardiac or vascular complications of the illness.

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Lyme Disease

LYME DISEASE is caused by a spirochete that can be found in blood, skin lesion or cerebrospinal fluid specimens. It has also been isolated from the nymphal and adult forms of its vector, *Ixodes dammini*. In California and Oregon, *Ixodes pacificus* ticks have been implicated. Cases have been widely distributed over the United States, Europe and the Far East.

Lyme disease typically begins in the summer with a unique skin lesion, erythema chronicum migrans, which usually lasts about three weeks, beginning as a red macule or papule that expands to form a large ring with central clearing. Annular lesions usually range from 6 to 52 cm in diameter; they may be absent or number more than 20 and last from three days to eight weeks. The rash may be accompanied by fever, headache, stiff neck, myalgias, arthralgias, malaise, fatigue or moderate lymphadenopathy. Weeks or months later, aseptic meningitis, meningoencephalitis, cranial neuropathies especially of the facial nerve, myelitis, migratory musculoskeletal pain, monoarticular or oligoarticular arthritis, myocarditis or atrioventricular node block may develop in certain patients. Radiculoneuritis may occur that is indistinguishable from brachial plexus neuritis. A Guillain-Barré-like syndrome, atypical in that it may show a cerebrospinal fluid pleocytosis in the range of 35 to 120 leukocytes, with 60% to 100% as mononuclear cells, has been described. Lymphocytic meningoradiculitis (Bannwarth's syndrome) has been associated serologically with Lyme disease. Serum cryoprecipitates, raised serum IgM levels and elevated erythrocyte sedimentation rates may occur.

Penicillin or tetracycline given for ten days can successfully treat the early phases of the disease when rash is present and can prevent, or at least ameliorate, the subsequent arthritic, neurologic or cardiac disorders.

Diagnosis can be confirmed by the finding of raised levels of IgG or IgM antibodies to the spirochete using indirect immunofluorescence, or by isolating the spirochete. These tests are presently of limited availability except through state departments of health, so local laboratory personnel should be alerted to the specific diagnostic concern when considering this disease.

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Pneumococcal Polysaccharide Immunization

IMMUNIZATION WITH pneumococcal polysaccharide vaccine is a safe and effective means of increasing a specific antibody and providing protection against overwhelming infection from *Streptococcus pneumoniae*. It is generally agreed, however, that immunization should be restricted to specific populations at high risk for infection.

Those patients who should be immunized include persons with increased susceptibility to infection but in whom a normal antibody response can develop following immunization. Specific diagnoses include sickle cell disease, splenectomy following trauma or for hematologic disorders, aging (that is, older than 55 years of age), complement disorders and nephrotic syndrome. Patients who might benefit from immunization but who have an impaired antibody response include those who have Hodgkin's disease with splenectomy, multiple myeloma and other malignant disorders. Many patients who are susceptible to overwhelming infection have severely impaired immunity and would not be expected to benefit from immunization. These include patients who have hypogammaglobulinemia and severe abnormalities of both T-cell and B-cell immunity. Patients with chronic lung disease who do not have an increased susceptibility to *S pneumoniae* infection, such as those who have asthma or cystic fibrosis, should not be immunized.

Immunization should be given to children 2 years

of age or older, as the antibody response to pneumococcal polysaccharide is significantly reduced in infants. However, selected patients who are at high risk for *S pneumoniae* infection or who have repeated infections may receive some benefit from immunization given at 9 months of age and followed by a booster at 2 years. In children older than 2 years of age and in adults, booster immunization should not be given before three to four years following primary immunization, as significant local and systemic reactions may occur. Recent evidence indicates that antibody levels to pneumococcal polysaccharide are reduced four years following primary immunization in older children and adults.

Several pneumococcal polysaccharide preparations are available for immunization. All contain 23 individual polysaccharides that have the ability to protect against 90% to 95% of *S pneumoniae* organisms that may cause infection. Immunization is given as a single subcutaneous dose of 0.5 ml. Local reactions, which are infrequent, consist of pain and swelling at the site of infection. Rarely, patients may experience more extensive swelling of the arm or systemic reactions such as fever, or both. Severe systemic reactions are extremely rare.

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α-Fetoprotein Screening

α-FETOPROTEIN is a serum protein that is normally found only in fetuses and is excreted via fetal urine into amniotic fluid. It probably reaches maternal serum via the placenta and possibly across the fetal membranes. Certain fetal abnormalities including surface defects allow fetal fluids containing high levels of α-fetoprotein to leak into amniotic fluid. Serum α-fetoprotein levels never exceed about 10 ng per ml in normal adults except in pregnancy.

Anencephaly, spina bifida and encephalocele, collectively called neural tube defects, are relatively common causes of fetal and infant death and survivors are often seriously handicapped. The incidence in the western United States is between 1 and 2 per 1,000 births. About 90% of infants with neural tube defects are born to couples who do not have a family history of this disorder and can be identified prenatally only by screening all pregnancies.

Several pilot studies confirm that at least 90% of cases of anencephaly and 80% of cases of open spina bifida can be identified prenatally by measuring α-fetoprotein levels in maternal serum. Laboratories involved in screening should have established their own normal α-fetoprotein range for each week of pregnancy from 15 through 22 weeks. Accurate gestational dates are essential, as serum α-fetoprotein levels increase by about 10% to 15% in each week over this period. Most protocols recommend a second blood test if the serum α-fetoprotein level exceeds the 95th percentile, followed by a sonogram if this is also elevated. In about half of these cases, a high serum α-fetoprotein level is explained by the presence of twins, incorrect gestational dates and, occasionally, fetal demise. Amniocentesis is recommended for the remainder of cases (about 1% to 2%) after a detailed ultrasound examination of the entire fetus. α-Fetoprotein assay of amniotic fluid should be combined with acetylcholinesterase gel electrophoresis, and repeated ultrasound examinations and biochemical testing may be necessary to confirm the diagnosis.

Maternal serum α-fetoprotein screening will identify a group of women at risk for an abnormal fetus. The ultimate diagnosis requires both sonography by experienced personnel and biochemical studies of amniotic fluid. The risk for a case of open neural tube defect after one elevated serum α-fetoprotein level is 1 in 50 and 1 in 15 at amniocentesis. The chance of terminating the pregnancy of a normal fetus is exceedingly small (less than 1 per 100,000) providing the protocol described is followed. Of women without fetal abnormalities, 80% can anticipate a normal pregnancy outcome, but there is about a fourfold increased risk of late pregnancy problems, such as fetal death and low birth weight. The early identification of twins and higher risk pregnancies are positive aspects, while anxiety generated by an abnormal serum value and an increased number of sonographies and amniocenteses are negative aspects of maternal screening.

It is anticipated that maternal serum α-fetoprotein screening will become a more generally available voluntary pregnancy test in the near future. This will require the coordination of laboratories and prenatal services, as well as educational programs and counseling services.

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Growth Hormone Therapy

FOR ALMOST three decades pituitary-derived human growth hormone has proved an effective treatment for children with absent or low growth hormone concentrations measured by radioimmunoassay of serum after stimuli by growth hormone secretagogues. Low plasma concentrations of somatomedins, growth hormone-dependent peptides believed to directly stimulate growth, are characteristic of such patients. Five years ago cases of short, poorly growing children with normal immunoreactive growth hormone and low somatomedin concentrations were described. Because they grew and their somatomedin levels rose when exogenous growth hormone was administered, they were considered to produce biologically inactive growth hormone that was still reactive in standard growth hormone radioimmunoas-